MACULAR HOLE AND ITS SURGERY

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INTRODUCTION

A macular hole is an anatomical opening or dehiscence in the fovea. It affects 33 of every 10,000 individuals over the age of 55[1]. Patients usually present with a painless disturbance in central vision and reduced visual acuity. Macular holes were previously considered untreatable until surgery was shown to be effective in the 1990's. In the decade that followed, improved surgical closure rates have been reported as a result of greater experience and newer techniques.

This paper reviews the causes of macular hole formation, its clinical presentation and staging. Existing treatment options as well as experimental techniques are also explored.

ANATOMY OF THE MACULA

The retina is the innermost nervous layer of the eye where images are formed by the eye's optical system. It consists of an inner neurosensory layer and an outer pigmented layer – the retinal pigment epithelium. Incoming light rays are focused on the retina and converted to nerve impulses that are then conducted along the visual system to the brain for higher cortical processing.

The macula is an oval area at the posterior pole of the globe measuring approximately 4.5 mm in diameter. It is concerned with central vision. The centre of the macula is a depressed area 1.5 mm in diameter called the fovea. The retina is thinnest at this point. There are no blood vessels at the centre of the fovea (the foveal avascular zone) and the photoreceptors rely on the underlying choriocapillaris for nutritional support. The floor of the fovea is called the foveola and consists almost entirely of a high concentration of cone photoreceptors. The nerve cells and fibers of the inner retina are displaced to the sides, which accounts for the thinness of the retina at the fovea. This structural arrangement allows incoming light rays to have the most direct access to the photoreceptors. Because the macula subserves the most acute vision, a full-thickness defect at the macula will manifest as a central visual defect and a reduction in visual acuity.

Causes

Macular holes are usually idiopathic. They may also be secondary to blunt trauma, high myopia or solar retinopathy.

It is believed that idiopathic macular holes begin with an initial dehiscence of the umbo and minimal loss of photoreceptors[2,3]. This may be caused by spontaneous, focal contraction of the vitreous at the fovea resulting in tangential, circumferential or anteroposterior traction which elevates the retina[4]. Eventually, further contraction and condensation of the vitreous may progress to a full-thickness macular hole.

Blunt trauma may result in commotio retinae that subsequently progresses to cystoid macular oedema and macular hole formation. Macular holes may also result from vitreous traction that later develops as a result of the trauma.

High myopia is associated with macular hole formation and may progress to a localised retinal detachment at the posterior pole.

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Staging

Macular holes are staged according to the classification proposed by Gass in 1995[2]. They usually progress through the various stages over a period of weeks to months.

Stage 1 holes are not full-thickness holes. An impending hole (Stage 1a) is characterised by a flattening of the umbo (loss of foveal depression) and a central yellow spot at the fovea. In occult holes (Stage 1b), there is a yellow ring at the macula.

Stage 2 holes are characterised by a full thickness hole with a diameter of less than 400 mm.

Stage 3 holes are full thickness holes with a diameter greater than 400 mm.

In Stage 4 holes, in addition to a full thickness hole greater than 400 mm in diameter, there is a Weiss ring in front of the optic nerve head. The Weiss ring is caused by posterior vitreous separation.

Clinical presentation

Patients with Stage 1 macular holes usually do not present clinically. They do not have a full-thickness retinal defect and therefore experience minimal reduction in visual acuity. Sometimes, even the more advanced holes may be detected incidentally during a routine ocular examination for reasons such as diabetic retinopathy screening. Patients commonly seek medical attention only when visual symptoms develop. These symptoms are sometimes discovered by chance when the better-seeing eye happens to be covered.

Patients complain of a painless loss of central vision (central scotoma) which may be associated with distortion of images (metamorphopsia) and reduction of visual acuity.

The diagnosis is made by fundal examination after pupillary dilatation. A round, punched-out area is seen at the macula, surrounded by a shallow rim of retinal detachment. The punched-out area has a darker color than the surrounding retina (Figure 1).

The Watzke-Allen test may be useful in determining whether a macular hole is full-thickness. A narrow slit beam is projected onto the macula across the suspected hole. If it is a pseudohole, the patient will report that the beam is either unbroken or has mild distortion or indentation of a portion of the beam. If it is a full-thickness hole, the patient will usually notice a break in the beam of light.

Investigations

History and clinical examination are usually sufficient for the diagnosis of a macular hole. A fundal fluorescein angiogram (FFA) can be performed in cases of uncertainty or for the purpose of establishing an objective record. The angiogram shows an area of hyperfluorescence which corresponds to the macular hole. The hyperfluorescence is due to a 'window defect' in the retinal pigment epithelium, which results in the unmasking of the choroidal vasculature, which lights up in the FFA.

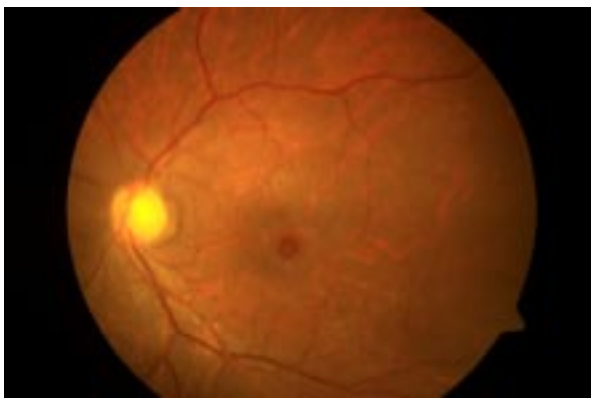

Figure 1: A full-thickness macular hole. The hole appears as a round, punched-out lesion at the centre of the macula. Visual acuity is 6/60.

Treatment of macular holes

Previously, no treatment was available for macular holes until a landmark paper published by Kelly and Wendel in 1991[5], established a successful surgical technique that resulted in anatomical closure. In the decade that followed, increased surgical experience and improved techniques have resulted in increasingly successful closure rates, from the initial reported rate of 58% to rates of over 90%.

The issues to consider in the management of macular holes are:

- Is surgery necessary?
- If surgery is performed, does adjuvant therapy improve the outcome?
- What are the potential risks and complications?

Clinical trials have demonstrated that patients with macular holes of Stage 2 or greater (full-thickness macular holes) benefited from surgery[6-8]. There were significantly higher rates of anatomical closure and improvement in visual acuity compared to patients who were treated conservatively. However, one study showed no benefit in performing surgery for Stage 1 macular holes compared to observation[9]. It should be remembered that most patients with Stage 1 holes do not present clinically.

Surgical treatment

The surgical treatment for macular holes is a three-port pars plana vitrectomy. This is the technique originally described by Kelly and Wendel[5].

Briefly, three ports are inserted at the pars plana (part of the ciliary body) to allow intraocular access for instruments, including a vitrector (vitreous cutter), a light source (endoillumination) and infusion. The vitreous is removed by the vitrector and the posterior hyaloid is detached from the macula by the vitrector or a soft tipped canula. This crucial step is confirmed by the appearance of a Weiss ring. The vitrectomy is then completed by trimming the peripheral vitreous.

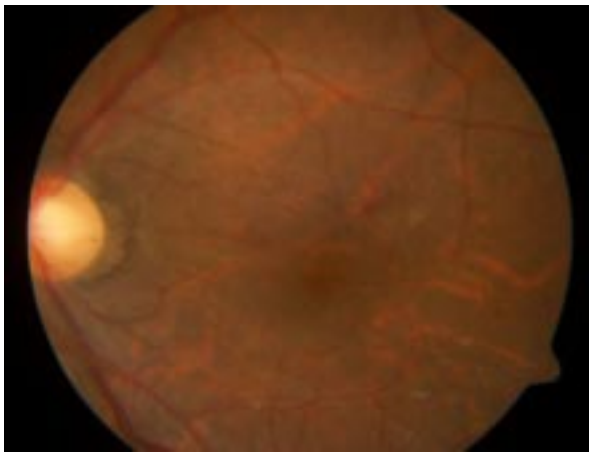

Figure 2: Same eye as Figure 1 four weeks following pars plana vitrectomy, intravitreal gas injection and post-operative face-down head positioning. The fundus photograph shows closure of the macular hole. The patient's visual acuity has improved to 6/24.

Following the vitrectomy, the peripheral retina is inspected for pathology. In particular, retinal tears may occur during the vitrectomy due to vitreo-retinal traction. When this happens, the retinal tears have to be treated with endolaser photocoagulation or cryotherapy.

Finally, the fluid in the vitreous cavity is replaced by a gas. Different types of gases have been used by various surgeons, including perfluoropropane (C_3F_8), sulphur hexafluoride (SF_6) and filtered room air. The gas bubble spontaneously disappears over several days to weeks, the exact duration depending on the gas used. The outcome following macular hole surgery is shown in Figure 2.

After surgery, patients are required to maintain a strict face-down head position (Figure 3) as much as possible for the next two weeks so that the macular hole is uppermost in the eye. This allows the floatation force of the gas to tamponade the macular hole. A special "pillow" called a vitrectomy

Figure 3: The post-operative face-down head posture keeps the macular hole uppermost in the eye and allows the intravitreal gas (or silicone oil) to float up against the hole. This internal tamponade helps to close the hole. The patient may maintain this head posture while lying prone, sitting or even standing. Postoperative head positioning is usually performed for about two weeks. However, patients can take short breaks to exercise their necks and limbs.

face support, is available to assist them in maintaining the posture. This allows them to posture comfortably while maintaining the head position over the two weeks. While the head is looking down, the patient can drink using a straw, read with the opposite eye or watch television using a mirror inclined at about forty-five degrees to the floor.

Although compliance with posturing is essential, it is equally important for patients to exercise and move their limbs from time to time to prevent stiffness and spasm. Very rarely, thromboembolism may occur in patients who are posturing without adequate movement, especially if they have thrombophilia[10].

Intravitreal silicone oil may be used instead of gas for patients who are unable to maintain the appropriate head posture for reasons such as stroke. It is also useful in patients who are required to travel by air soon after the surgery. The use of silicone oil minimises the inconvenience to the patient and a closure rate of 80% has been reported[11]. However, because silicone oil does not disappear spontaneously, a significant disadvantage is that patients will require another intraocular procedure to remove the silicone oil at a later date.

Adjuvant Therapy

Adjuvant therapy, internal limiting membrane (ILM) peeling and laser photocoagulation have all been used, in addition to pars plana vitrectomy, in attempts to improve the success rate of macular hole surgery.

A variety of adjuvants have been tried in macular hole surgery. These include transforming growth factor b_2 (TGF-b_2), autologous platelet concentrate (APC), autologous serum, thrombin, plasmin and thrombin-activated fibrinogen. Surgery relieves traction on the macula, after which the hole is closed by the formation of a glial scar that helps bridge the defect in the neurosensory retina, reattaching it to the edges of the hole. The use of adjuvants is based on the premise that they may help stimulate fibrosis and wound repair, which is necessary for hole closure. For example, TGF-b_2 is a growth factor that is instrumental in fibrosis and wound repair, while platelets contain many growth factors in the alpha granules with potential healing activity[12].

Currently, available clinical evidence does not suggest any definite additional benefits of adjuvant therapy when compared to surgery without the use of adjuvants[6,12-16]. It should be pointed out that at this time, only a few double-blind, randomised clinical trials on the effectiveness of adjuvant therapy have been performed. Most of the remaining data come from uncontrolled case series and cohort studies. More detailed studies on the use of adjuvants are required before their effectiveness can be determined.

Complications

The complications associated with macular hole surgery are similar to those encountered in other types of vitreous surgery[6].

Over 75% of patients will develop cataracts

after the surgery. Retinal detachment may occur and a frequency of between 2 to 11% has been reported. Patients should be aware that there is a 2 to 10% risk of recurrence which may require additional surgery.

Visual field defects may occasionally develop and is believed to be due to trauma or dehydration of the nerve fiber layer during the surgery.

Rarer complications include endophthalmitis, uveitis, raised intraocular pressure (glaucoma) and retinal pigment epithelium changes.

Prognosis

The prognosis for macular holes is excellent and most patients can be successfully treated with surgery, resulting in anatomical closure and improvement in visual acuity.

Patients should be advised that about 10% of people with idiopathic macular holes may subsequently develop a macular hole in the fellow eye[17,18].

CONCLUSION

A macular hole is a treatable condition. Patients with macular holes of Stage 2 or greater are likely to benefit from surgery. Although some studies involving adjuvants have yielded promising results, at present, there is still no definite evidence that the use of adjuvants improves the surgical outcome.

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